

MALIGNANT PHEOCHROMOCYTOMA: ABOUT SEVEN OBSERVATIONS:EP -50

INTRODUCTION

Malignant pheochromocytomas are rare tumors, developed at the medulla and paraganglia .. Their diagnosis is established by the presence of metastases of organs devoid of chromaffin tissue or by the appearance of neoplastic recurrence . They are characterized by a morbidity and mortality due to the effects of uncontrolled and important hypersecretion cathécolaminergique and resistance to conventional cancer treatments

AIM

Report the observations of seven cases observed in 26 years

MATERIALS AND METHODS

This is a retrospective study of malignant pheochromocytomas 's cases hospitalized in our service. we analyzed the clinical, biological, radiological, treatment and evolutionary of these neoplasias

RESULTS

Seven patients with malignant pheochromocytomas were treated in our department.

3 were male and 4 female. their ages ranged between 17 and 45 years old. The circumstances discoveries were adrenergic signs and severe hypertension in all cases .

Hormonal balance showed very high levels of metanephrine: 20 ± 1.4 (16- 22)

and CT + MRI showed a large and characteristic adrenal mass : 12.6 ± 1.2 cm (14- 9.6). Malignancy was confirmed by the presence of metastasis at diagnosis in 5 cases and in the developments in the rest of the cases

All patients have been operated. The surgery was complete in 5 cases and partial in 2 cases

Chemotherapy was conducted in all cases supplemented by therapeutic cures MIBG in three cases

Evolution was marked a year and a half after (8 months- 4 years) after surgery and chemotherapy by a normalization of blood pressure, methoxylated derivatives in 5patients.

Two deaths occurred after the first course of chemotherapy and three deaths after a mean of 2 years of recurrence and secondary metastases

DISCUSSION AND CONCLUSION

Malignant pheochromocytoma is even rarer. It represents about 10% of cases. Its diagnosis is difficult because there are no histological criteria of certainty. Only a local recurrence and metastases or the presence of chromaffin tissue in organs usually devoid of can confirm the diagnosis

The clinical presentations are varied and depend on the secretory profile. Tumors secreting norepinephrine mainly cause vasoconstriction and diastolic hypertension. When the adrenaline is predominant, we observe tachycardia, systolic hypertension with orthostatic hypotension and risk of major hypotensive access, or non-cardiogenic pulmonary edema. If preferential or exclusive secretion of dopamine, there is usually no arterial hypertension and the diagnosis is often suspected in atypical manifestations or fortuitous discovery

Therapeutic management is very difficult. It is based on surgical resection whenever possible including cases of metastases. Indeed partial surgery increases survival. In extensive and inoperable forms, therapeutic options are limited by the low sensitivity of tumor cells with chemotherapy and radiotherapy . Apart from symptomatic medical treatment of adrenergic hypersecretion by alpha blockers and beta blockers, MIBG labeled with iodine 131 is, at present, the main treatment option for fixing the tracer tumor sites

Surgery of malignant pheochromocytoma, even at the stage metastatic must be done because it increases survival. Chemo radiotherapy and MIBG constitute an important part of the therapeutic arsenal and should be considered secondary. The management of these patients requires a multidisciplinary collaboration involving endocrinologists, surgeons, oncologists, nuclear physicians and pathologists. The prognosis remains pejorative.

